107 Hair

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Definition

The human pelage, compared to that of other mammals, is a meager thatch of limited physiologic worth. Its psychologic value is incalculable. Consider the extreme example of the patient who resists cancer chemotherapy because it will produce temporary hair loss. Life, or the quality of life, is being influenced by what could be cynically dismissed as useless ornamentation. The importance to such patients is acknowledged by the considerable medical efforts that may be directed at preventing the loss to help ease the travails of unpleasant therapy. Because hair is the source of such focused attention, abnormalities are quickly perceived by the patient. For the physician, there is the opportunity for early detection of occult systemic disease.

Basic Science

The production of hair requires considerable metabolic effort. On the scalp alone, where 100,000 hairs grow at the daily rate of 0.3 to 0.4 mm, it roughly translates to the equivalent of producing a single tough, protein filament approximately 40 m in length! This requires fine-tuned nutritional, vascular, and hormonal interactions, and it is reasonable to expect that a variety of systemic diseases will produce abnormalities in hair growth. Additional traumas are the result of the environment, and a variety of physical and chemical assaults visited on the hair in the name of grooming, according to the current dictates of fashion.

Hair greatly resembles the epidermis from which it is derived. An active matrix of growing cells at the dermo–epidermal interface differentiates to produce a mass of dead cells composed primarily of the tough protein, keratin. In place of a sheetlike stratum corneum or protective horny layer, the cells are packed radially in longitudinal array in the central cortex and invested with a circular band of overlapping cells, the cuticle, which binds the mass into a cohesive package. Like a good cigar, the wrapping must be perfect or the contents will unravel, splay, and fragment. As in the epidermis, pigment is incorporated into the keratinizing cells by melanocytes, right at the matrix. The growing end of the hair is deep within the skin near the subcutaneous fat—in the case of the scalp, about 3.0 mm below the surface. Most of the fiber, though connected to the relatively minuscule growing portion, is a passive, dead structure, which is simply pushed along and out. There is no exchange of vital juices or controlling signals to the productive end. The external fiber can thus be cut, bleached, curled, and straightened without peril to growth, even though the exposed hair itself may be obliterated.

Hair differs from the epidermis in one very important way. While skin is continuously growing and replacing its protective horny layer, hair grows in cycles. The obvious anatomic differences of eyebrow hair versus scalp hair, for example, reflects their varying growth, rest, and replacement phases. Tresses extending from crown to rump, about 1 m, necessarily require 1000 days in the active growth, or *anagen*, phase. About 90% of scalp hairs are in anagen daily, while the other 10% are being shed and replaced, the so-called *telogen* phase. Everyone can, with gentle traction, dislodge these telogen hairs, which are easily recognized by their characteristic naked, white, or hypopigmented spheroid terminus. Patients routinely consider these the "roots," but they are in reality the totally keratinized portion of the hair that is being pushed out by an emerging new hair, since the regenerative matrix remains deep within its protected sanctuary. Anagen hairs must be traumatically plucked out of the skin. In doing so they bring along their anchoring sheaths and appear to have gelatinous casts surrounding the dark, pigmented, plastic cylinder of living cells that have been torn from the subadjacent nurturing tissue. Occasionally, the acetabulum-like depression, which had surrounded the dermal papilla, remains visible. Because replacement of telogen hairs proceeds regularly, some amounts of daily shedding are to be expected. Although up to 100 hairs may fall from the scalp, there is actually no net loss.

Clinical Significance

Patients typically seek medical care for problems of hair loss or excessive hair growth. These complaints, which may have little physiologic effect, can cause considerable psychologic distress to the patient.

By far, the most common complaints and signs of abnormal hair growth are associated with its loss. *Alopecia* means hair loss or baldness. The term is said to be derived from the Greek word for fox, "an animal that commonly suffers a mange" which causes hair loss.

Unwanted excess hair is as cosmetically distressing to the patient as alopecia. *Hypertrichosis* is the term applied to excess hair growth on any part of the body, while *hirsutism* is meant to be limited to androgen-induced, coarse, dark growth in special areas, such as the beard.

Technique

The history should include careful questioning for illnesses or trauma that might explain complaints of hair loss. Such seemingly ordinary practices as the use of a curling iron or hair dyes or bleaches can damage the hair shaft. In the case of excessive hair growth, the history should look for genetic, physiologic, or metabolic clues to the etiology.

A complete examination of the scalp and other body hair, as indicated, should be performed to assess bald spots and thinning; evidence of trauma to the scalp or hair shaft; parasites; pattern, location, and amount of excess hair

growth; and changes in the pigmentation, color, and texture of the hair.

In alopecia, when the complaint is excessive loss rather than bald spots, a global view of the patient seen at an initial visit may be misleading. It has been estimated that as much as 25% of scalp hair may be diffusely and acutely missing without the defect being apparent to the examiner. Comparison with a recent photograph may be possible. If attempts are made to have the patient determine whether "excessive loss" is real, hairs may be collected and counted. This is best accomplished by a thorough daily shampoo to avoid the inaccuracy of an apparent increase in fallout due to accumulation between washings.

If the condition is still in an active phase of loss, gentle finger traction will dislodge the evidence, and the examiner will be able to look at the proximal ends to help determine the cause of the effluvium. Are there typical telogen hairs? Are the shafts tapered? Are there broken or fractured shafts? The answer can be usually seen with the naked eye, although a magnifying lens or low-power light microscope might be needed. The texture of the individual fibers and the manner of light reflection along their length are estimates of qualitative changes to the hair.

Careful observation is made to determine whether the hair loss is diffuse throughout the scalp or limited to circumscribed areas. In the bald spots, the scalp is examined for signs of scarring or evidence of surviving follicles. The more common alopecias can then be conveniently categorized into distinct groups and the appropriate diagnosis determined (Table 107.1).

Noncicatricial Alopecia, Patterned Type

MALE AND FEMALE PATTERNED ALOPECIA

The typical baldness that accompanies aging in most men is so common as to be considered physiologic. It proceeds after puberty with a frontotemporal recession, followed by a thinning at the vertex, and the subsequent joining of both parts to form a denuded horseshoe-shaped pate surrounded by a surviving peripheral fringe. In the active phases of loss there is increased shedding of telogen hairs as greater numbers of follicles go through progressively shorter growth cycles. This is mirrored in the gradual development of smaller, thinner, less pigmented hairs in the balding areas. Finally, all that remains is a barely visible fuzz. Except that a minimum amount of androgenic hormonal activity is required, the process may be conveniently viewed as an aging process of individual follicles in a susceptible locale. The fact that surviving hairs may be successfully transplanted from the parietal scalp into the bald areas is eloquent testimony to the adequacy of the underlying vascular and nutritional milieu.

In women, frontocentral balding, otherwise similar to the variety described in men, occurs in predisposed patients at a slower pace, with advancing age. Estrogen loss after menopause may allow the androgenic influence to hold sway. Early, severe loss, particularly with associated signs of hirsutism, acne, and menstrual abnormalities, should lead to a search for excessive androgens whether from endogenous ovarian or adrenal overproduction, or exogenous sources such as the progestogen oral contraceptives.

Table 107.1
Classification of Alopecia

Noncicatricial alopecia
Patterned
Male and female patterned alopecia
Traumatic alopecia
Alopecia areata (A. totalis, A. universalis)
Tinea capitis
Diffuse
Telogen effluvium
Anagen alopecia
Congenital dystrophies
Endocrinopathies
Cicatricial alopecia
Patterned
Discoid lupus erythematosus
Morphea (localized scleroderma)
Hot comb alopecia
Tumors
Diffuse
Deep tissue trauma, infection

TRAUMATIC ALOPECIA

The most common hair problems of women stem from damage caused by styling and grooming. The traction from "pony tail" fashions, tight "corn row" braidings, the pressure from sleeping with rollers in the hair, etc., all produce hair loss in specific geometric patterns that correspond to the site of prolonged trauma. A counterpart is seen in *trichotillomania* where habitual pulling and twirling the hair produce less regular patterns. This nervous habit is not uncommon in children but portends a more serious psychiatric problem in an adult. In the traction alopecias, short, uneven regrowing or broken hairs remain and help point to the diagnosis.

The chemical treatments of bleaching, coloring, waving, and straightening hairs, if too harsh, too prolonged, or too frequent will produce cumulative damage to the fibers. Their structural integrity destroyed, the hairs simply break at the weakest points. The fragmented ends are visible in the many pieces easily removed by rubbing, washing, or combing the hair. The remaining hairs are usually of the same general length but are quite uneven, resulting in a ragged-looking, unintentional "crew cut."

ALOPECIA AREATA

The explosive onset of circles of baldness of any hair-bearing part of the body announces alopecia areata. The process appears in a random and unpredictable fashion and may proceed to loss of all scalp hair, *alopecia totalis*, or from the entire body, *alopecia universalis*. Circumscribed scalp loss is the variety most commonly seen. The scalp in the affected sites appears remarkably normal and serene, effectively masking the severe inflammatory process in the depths of the hair matrix. Surviving hairs, if present, are invariably depigmented. Reports of hair turning white overnight represent selective loss of pigmented hairs in alopecia totalis. No physiologic mechanism exists that will enable hair to alter its color suddenly. Alopecia areata may also be associated with vitiligo, and both conditions are thought to be autoimmune processes. There also appears to be an association between alopecia areata and organ-specific endocrine disorders, particularly thyroid disease.

TINEA CAPITIS

Within the past two decades the character of "ringworm of the scalp" has changed dramatically. Classic tinea capitis

occurred as single or multiple annular patches of dull, scaly alopecia containing the small stubble of many broken hairs. The majority of causative organisms produced metabolites that fluoresced yellow-green under long-wave ultraviolet (Wood's) light. Diagnosis was confirmed by digesting a hair remnant with potassium hydroxide solution and microscopically viewing the fungal spores clustered about the hair. The relative ease of diagnosis and the development of effective therapy have eliminated the "classic" genera, but have selectively allowed other fungi to flourish. The tineas seen currently do not fluoresce. The infections are deep within the follicle and produce the so-called black dot ringworm. Here the hairs are at or below the surface and are very difficult to obtain for direct microscopic examination. The clinical presentations are not always regular circles, but may be irregular and inflammatory. They also produce scarring. The current tineas of the scalp require a high index of suspicion and energetic laboratory investigation including cultures and biopsy to establish the diagnosis.

Noncicatricial Alopecia, Diffuse Type

In early stages, diffuse loss is only noticed as increased hair fallout, which later shows as general thinning and a clearly visible underlying scalp. The not-too-apparent early fallout is usually confirmed either by the tug or traction test, already described, or by having the patient save and count the daily hair loss.

TELOGEN EFFLUVIUM

A wide variety of conditions and factors may precipitate the temporary shedding of increased numbers of otherwise normal telogen hairs. There is a common tendency to attribute such findings to the patient's "nerves" based on anecdotal reports; such associations should be viewed with skepticism.

In *postpartum effluvium,* hormonal influences during the last trimester of pregnancy appear to inhibit the normal shedding of hair so that some women notice a thicker, more luxuriant growth. In the postnatal period the condition reverses, and, within weeks to months after delivery, an accelerated loss of telogen hairs occurs. This delayed shedding causes great concern, but the hair merely returns to its prepregnancy status. The alarmed patients require assurance that the event is temporary and that there will be no real loss of hair.

Febrile-toxic effluvium is another form of noncicatricial alopecia. Severe illnesses, particularly infectious diseases marked by high fevers, may have an associated telogen effluvium. The uncommon "moth-eaten" alopecia of syphilis is probably the best-known example. Complete restitution of hair follows appropriate therapy of the underlying disease. Alopecia of this variety is also seen in patients who undertake rapid weight reduction, starvation diets.

Drug-associated hair loss can result from a wide variety of agents. The progestogen oral contraceptives, high doses of vitamin A, the newer therapeutic retinoids, cimetidine, beta-blocking agents, and nonsteroidal anti-inflammatory agents, among others, can produce temporary telogen hair loss. The coumadin and heparin anticoagulants appear to have a special proclivity for producing a telogen effluvium, which rights itself even with continued therapy.

ANAGEN ALOPECIA

Drugs and other poisons with selectively toxic effects on cells having high metabolic and mitotic activity, like cancer cells, will affect the hair matrix. Most cancer chemotherapeutic agents, x-rays, and heavy metals, such as arsenic, lead, thallium, and bismuth, abruptly halt the production of hair. This produces a severely tapered narrowing of the fiber, which then breaks at the constriction. The arrow-like ends are diagnostic. Depending on the dose and duration, a diffuse thinning, moth-eaten appearance or complete denudation may result. In therapeutic situations, either vascular compression or severe hypothermia is utilized to prevent the local effect on the scalp, while the chemotherapeutic agent seeks out the target cancer. Otherwise, the hairs will regrow when the drugs are stopped.

CONGENITAL DYSTROPHIES

Rare inheritable conditions produce abnormal hairs and diffuse, nonscarring alopecias. A dominantly inherited disorder, *monilethrix,* shows alternating, regular constrictions along the length of the fiber. The hair breaks at the weakened isthmus and either becomes impacted within the follicles or grows as very short, irregular tufts above the scalp. "Moth eaten" is an apt description. Some patients have associated aminoacidurias, suggesting a defect in the biochemical synthesis of the hair. This has also been seen in *trichorrhexis nodosa,* a rare recessively inherited disease. Trichorrhexis nodosa, however, is more commonly encountered in sporadic cases without any biochemical abnormalities. The nonfamilial form is due to mechanical and environmental damage to the hair and is analogous to the "frizzies" or split ends found at the distal end of hairs that have been allowed to grow for long periods without cutting. Both the familial and nonfamilial varieties show the same structural defect in the cuticle that allows the inner cortical cells of the hair to splay out, looking much like two paint brushes thrust together, bristles to bristles. Clinically, the hairs show alternating patterns of reflected light that appear white at the defects and resemble so many nits or lice egg casings, studded along the length of the fiber. Breakage occurs at weakened points in a random fashion, producing yet another "moth-eaten" alopecia. Additional varieties of structural growth defects of hair accompany other rare congenital ectodermal defects in which sweat glands, nails, and teeth may also be abnormal.

ENDOCRINOPATHIES

Hair loss caused by the various endocrine disorders is probably more cited, and looked for, than seen. Aside from obvious virilizing adrenogenetic patterned alopecias, only the hair loss associated with severe hypothyroidism appears to be significant. In addition to generally dry skin, the hair is coarse, brittle, and sparse. Loss of the lateral half of the eyebrows, although found in hypothyroidism, is also present in secondary syphilis and leprosy.

CICATRICIAL ALOPECIAS

Permanent hair loss from scarring of the scalp is an uncommon clinical problem. The patterned alopecias occur more frequently than diffuse generalized scarring loss. *Discoid lupus erythematosus* may present with annular, atrophic, denuded lesions anywhere on the body, including the face and scalp. Early lesions will show an intense erythema and characteristic adherent white scale. When removed man-

ually, the scale shows spikes or projections, "carpet tack" defects, which represent the follicular hyperkeratosis associated with the condition. *Morphea* or *localized scleroderma* occurs as annular to linear patterns of hairless sclerotic tissue. In the scalp a particularly striking defect, appropriately labeled "coup de sabre" deformity, appears to cleave a straight longitudinal furrow that can extend down the forehead and onto the face.

Two scarring alopecias seen predominantly in black patients are distinctive. *Hot comb alopecia* occurs in middle-aged and elderly women and results from the chronic thermal damage of the hair-straightening procedure. The crown is primarily affected, sparing the peripheral scalp. The pattern develops as petrolatum, used to facilitate movement of the hot implement over the hairs, trickles down the hairs and gradually burns out the areas. The process is rarely total, so sparse surviving follicles, some bearing multiple hairs, are interspersed in the otherwise barren landscape. *Dissecting cellulitis of the scalp* is seen more commonly in black men. A chronic infection begins as a minimal folliculitis, usually at the nape of the neck, progresses to form deep abscesses, which then burrow and spread, undermining large areas of the scalp. Boggy purulent tunnels develop and ultimately severe scarring, with widespread, irregular swaths of alopecia.

Space-occupying *tumors*, benign and malignant, may produce circles of alopecia that superficially resemble alopecia areata. The *wen*, probably the most common tumor of the scalp, though devoid of hair, is paradoxically derived from the follicular tissues (tricholemmal cyst). These solitary or multiple cysts are superficial, freely movable, and painless. The *nevus sebaceus* is an organoid hamartoma of epithelial tissue seen in infants at birth. The plaque of alopecia usually present at the vertex is a yellowish, verrucous, infiltrated, round or oval tumor from one to several centimeters in diameter. Aside from the cosmetic defect, 10 to 40% of the tumors undergo neoplastic change, usually to basal cell carcinomas during the second and third decades.

Diffuse cicatricial alopecia implies catastrophic destruction of the deep scalp, the culmination of any severe process: infection, physical or chemical trauma, x-radiation, and so on.

Increased Hair Growth

Both hypertrichosis and hirsutism in women must always be judged in terms of the individual's familial and ethnic background. With age, after menopause, some degree of hirsutism is to be expected, purportedly due to diminished estrogen production and a relative increase in androgen stimulation of the follicles. In younger women, particularly if accompanied by menstrual irregularities, fertility problems, acne, alopecia, and clitoral enlargement, hirsutism should trigger a search for pituitary, adrenal, or ovarian dysfunction or tumors. As in the case of alopecia, a wide variety of exogenous drugs will cause increased hair growth (Table 107.2).

Table 107.2
Increased Hair Growth Due to Drugs

Hypertrichosis	Hirsutism
Corticosteroids	ACTH
Cyclosporin	Acetazolamide
Diazoxide	Danazol
Dilantin	Phenothiazines
Minoxidil	Testosterone
Penicillamine	
Psoralens	

A peculiar rare familial form of excess generalized downy hair growth known as *congenital hypertrichosis lanuginosa* accounts for the individuals exploited as "dog-faced boy" or "human werewolf" at circus sideshows. Also rare, but important as a sign of internal malignancy, is *acquired hypertrichosis lanuginosa*. This "malignant down" usually precedes the neoplasia and has been reported with a wide variety of malignancies.

A characteristic, gross hypertrichosis of the face and extremities occurs in *hepatic porphyria*, along with mottled pigmentary changes, increased skin fragility, and blistering.

Circumscribed areas of excessive hair growth may occur following chronic irritation of the skin, by rubbing, as under plaster casts, biting (nervous tic), and following burns and ionizing radiation. Congenital tufts of hair along the midline, such as the sacral "faun tail," may be associated with underlying spinal defects.

Other Conditions

Aside from the obvious quantifiable changes of increased or decreased hair growth, patients manifest, or complain of, changes in pigmentation, color, and texture. These alterations also require explanation and may be clues to underlying systemic disease. Detailed catalogues of the sometimes vivid, and sometimes subtle, variations in hair growth are available, and will point the perceptive clinician to the required investigation and therapeutic intervention.

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